Grisel Syndrome

A Review

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Abstract

Grisel syndrome is defined as atlanto-axial subluxation following infections of upper airway. These patients present with torticollis which is persistent. Radiology is normal in these patients. This is actually a sinister problem which should always be kept in mind before taking up a patient for adenotonsillectomy. Subclinical subluxation of atlanto-axial joint may become overt causing brain stem compression following adenotonsillectomy surgery. This actually occurs because the paraspinal muscles are relaxed (due to the use of muscle relaxants) during the procedure. The actual position of tonsillectomy (Rose position) aggravates this condition. This article attempts to explore this problem with a review of 10 year published literature on this subject.

Synonyms:

Non traumatic atlanto-axial subluxation, distension luxation, spontaneous hyperemic dislocation, inflammatory dislocation, torticollis nasopharyngien.

Introduction:

Subluxation of atlanto-axial joint occurs due to:

1. Congenital causes

2. Traumatic causes

3. Iatrogenic causes (following surgical procedures)

4. Due to infections (Grisel's syndrome)

Fielding and Hawkins[1] classified subluxation of atlanto-occipital joints into 4 types:

Type I:

Simple rotatory displacement without anterior shift. The transverse ligament is intact and the dens acts as the pivot point.

Type II:

Rotary and anterior displacement between 3-5 mm. The transverse ligament is injured in these patients, the opposite facet acts as the pivot point

Type III:

Rotatory and anterior displacement greater than 5 mm. Both lateral atlanto-axial joints are subluxed anteriorly. The transverse ligament and facet capsules are injured.

Type IV:

Posterior subluxation of both lateral atlanto-axial joints. This type of dislocation is rather rare. It is seen in adults with rheumatoid arthritis. There is always the presence of destruction of dens.

Grisel syndrome:

This condition was described by Grisel in 1930 [2]. It should be pointed out that this condition was originally reported by Bell in 1830 nearly one century earlier. This syndrome is characterised by atlanto-occipital dislocation / subluxation. Patients with this syndorme manifest with torticollis with history of upper airway infection. Patients with this syndrome classically present with:

Painful head tilt (Cook-Robin Look). The chin is turned towards one side while the neck is laterally flexed to the opposite side. These patients have limitations in neck mobility. Attempted corrective rotation causes pain.

Clinical Features:

1. Children are frequently affected because they are more prone for adenoid inflammation. Anatomically adenoid is located in the area drained by pharyngoverterbal plexus. Infection could travel through this plexus to involve the stabilizing ligaments of atlanto-axial joints.

2. Presence of spontaneous torticollis

3. Neck stiffness and painful neck movements

4. Associated rhinopharyngitis[2]

5. Secretory otitis media due to eustachean catarrh

This is actually a more dangerous complication of tonsillectomy

Examination of nasopharynx may show reduction in the size of nasopharynx due to anterior displacement of the arch of atlas. In fact the projection of atlas can be palpated.

Neurological examination could be normal in these patients. Rarely these patients may manifest with C2 radiculopathy, high cervical myelopathy. Sometimes sudden death have also been reported.

Neurological manifestations include:

1. Mild paraesthesia

2. Exaggerated tendon reflexes

3. Bladder dysfunction

4. Quadriplegia

5. Acute respiratory failure & death[3]

Voice change:

Sudden change in nasal resonance due to reduction in the size of nasopharynx caused by subluxation of atlanto-axial joint may be seen.

Pathophysiology:

Atlas is a cervical bone which is flat with slanting articular facets on the lateral masses. These

articular facets form synovial joints with corresponding anterior articular surfaces of axis. This Capsule is loose and just allows minimal rotation and minimal lateral displacement of atlanto axial joint. There are two stabilizers acting on atlanto axial joint.

Primary stabilizer:

This is the transverse ligament [4]. This ligament is attached to the lateral posterior portion of the anterior arch of atlas thus forming posterior support of odontoid process preventing excessive shift of C1 on C2.

Secondary stablizers:

Paired alar ligaments form the secondary stabilizers of this joint [4]. They help in preventing excessive rotation of C1 on C2.

Theories accounting for pathophysiology of Grisel's syndrome:

1. Numerous venous tributaries (pharyngo-vertebral veins) drain the posterosuperior portion of nasopharynx. These veins cross the prevertebral fascia and drain into periodontoidal plexus, which empties ultimately into the upper cervical epidural sinuses. Inflammation spreads to these stabilizing ligaments via the anastomosis between lymphatic vessels and pharyngo-vertebral veins. This causes inflammatory effusion and stretching of stabilizing ligaments leading on to subluxation.

2. Irritative contracture of suboccipital and paravertebral muscles due to cervical lymphadenitis due to nasopharyngeal infections may cause subluxation. In addition regional hyperemia due to inflammation may cause decalcification of the attachment of transverse ligament causing ligamentous laxity and subluxation [5].

3. Spasm of irritated neck muscles can causes distension of ligaments and subluxation of the atlanto-axial joint [6].

4. There is increased risk of subluxation of atlanto-axial joint in children with Down's syndrome because of laxity of ligaments arount this joint [7].

5. Excessive extension / hyperotation of head under anaesthesia during otolaryngological surgical procedures (adenotonsillectomy) can lead to subluxation of atlanto-axial joint because the stablizing effects of the muscles and ligaments around this joint are reduced during anaesthesia.

According to published literature substantial number of published cases followed / precipitated by surgery. Common surgical procedure implicated is adenotonsillectomy. A few patients have been documented to have developed atlanto-axial subluxation following mastoidectomy also[8].

Suggested radiological features:

In lateral view cervical radiographs the lateral masses of first cervical vertebra will not be superimposed and may be visible anterior to the odontoid process. In antero posterior view the spinous process of C2 will be deviated towards the side of chin deviation. This feature is known as the Sudeck sign. The normal atlanto dens interval in lateral radiographs of cervical spine is about 3 mm in adults and 4.5 mm in children. Increase in atlanto dens interval of more than 4.5 mm suggests subluxation of atlanto-axial joint[9] .

CT scan really clinches the diagnosis. According to Li and Pang CT images should be obtained in 3 different positions [10]. Best visualization of this area is possible only with a 3 D CT scan images.

Even though subluxation in Grisel syndrome occurs in atlanto-axial joint, a variant of Grisel syndrome has been reported. Martinez-Lage[11] reported a patient with features of Grisel syndrome with subluxation of C2-C3.

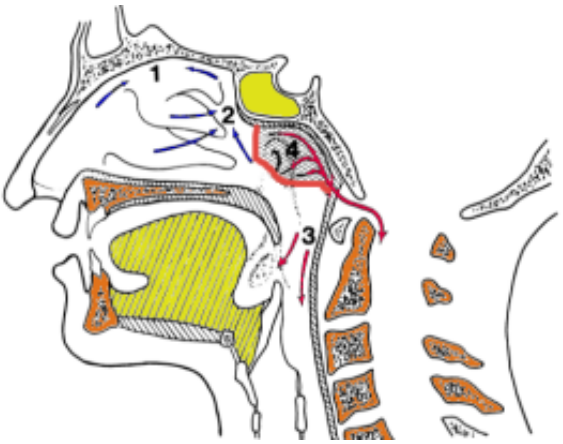

Diagram illustrating venous drainage of nasopharynx and cervical spine. 1. Venous drainage from upper part of lateral wall of nasal cavity goes into orbital veins via the anterior and posterior ethmoidal veins 2. Venous drainage from the lower part of lateral nasal wall and nasal septum drain into pterygoid plexus via the tributaries of sphenopalatine vein 3. Oropharynx drains into tonsillar tributaries of the maxillary vein and the posterior pharyngeal plexus 4. Pharyngo vertebral venous plexus drainage area

Management:

Treatment of infection:

Use of Intravenous antibiotics. Should be administered for 4-6 weeks

Pain management:

Use of antiinflammatory drugs

Traction

Type I Grisel syndrome: Soft collar

Type II Grisel syndrome: Hard collar

Type III Grisel syndrome: Halo fixation 13 . The concept of Halo vest was introduced in 1950. This vest attaches to the body by adjustable metal bars. It is attached to the patient's torso while stabilizing the neck. The major advantage of this brace is that this vest allows fixation of cervical spine while permitting the patient to stay mobile. The head is stabilised by 4 titanium pins which are inserted into the skull under local anesthesia. These pins are then attached to the brace which clothes the chest of the patient. Once the vest is in place radiographs of cervical spine should be taken to ensure that the cervical spines are adequately immobilized and repositioned. This vest should be used for a period not less than 3 months. Of course it has its own set of complications like infection, pin loosening, pin migration etc.

Type IV Grisel syndrome: Open fixation This is resorted to when neurological complications arise.

Avoiding atlanto-axial subluxation while performing otolaryngological surgical procedures:

1. Awareness of this syndrome

2. Avoidance of using unipolar cautery during tonsil surgeries

3. Avoid performing surgeries on patients with active upper respiratory infections / adeno tonsillitis

Figure showing Patterson's Collar

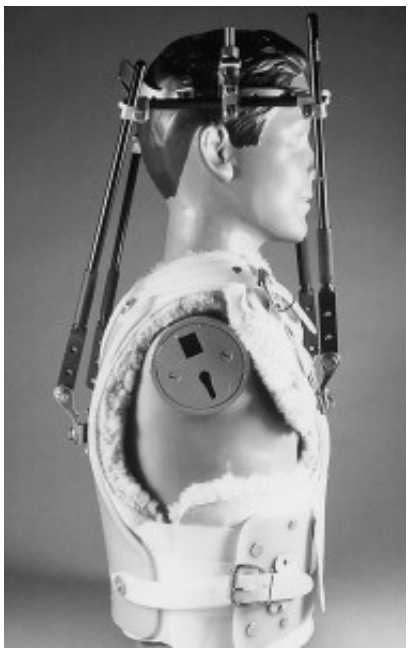

Figure showing Halo vest

Conclusion:

1. Grisel syndrome is commonly seen in children

2. This is a dangerous condition

3. Presence of torticollis following otolaryngological surgery / upper respiratory infections should arise suspicion of Grisel syndrome

4. Having identified this syndrome patient needs to be carefully observed, monitored and treated accordingly

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